Gynecologic Oncology Reports 9 (2014) 18–20

Contents lists available at ScienceDirect

Gynecologic Oncology Reports

journal homepage: www.elsevier.com/locate/gynor

Case Report

Umbilical endometriosis with giant degenerated uterine leiomyomas: A case report

Makiko Omori *, Tatsuyuki Ogawa, Masatoshi Nara, Akihiko Hashi, Shuji Hirata

Department of Obstetrics and Gynecology, Faculty of Medicine, University of Yamanashi, Yamanashi, Japan

ARTICLE INFO

Article history:
Received 31 March 2014
Accepted 27 April 2014
Available online 5 May 2014

Keywords:
Umbilicus
Endometriosis
Umbilical tumor
Uterine leiomyoma
Cystic degeneration
Magnetic resonance imaging

Introduction

Metastatic tumor of the umbilicus has been well known as a Sister Mary Joseph nodule (SMJN). The most common primary sites for metastases in women are reportedly the ovary (34%), endometrium (12%), colon/rectum (12%), stomach (9%), and pancreas (8%) (Dubreuil et al., 1998). SMJN has an extremely poor prognosis, and the differential diagnosis thus requires histological examination (Dubreuil et al., 1998). Nevertheless, umbilical tumors in women included endometriosis in 32.2%, benign primary tumor in 29.7%, metastatic tumor in 29.7%, and malignant primary tumor in 8.4% (Barrow, 1966). Umbilical endometriosis, one type of cutaneous endometriosis, is uncommon with an estimated incidence of 0.5–1.0% among all cases of endometriosis (Latcher, 1953). We report herein an unusual case of umbilical endometriosis and giant uterine leiomyoma with marked hydropic and cystic degeneration, in which magnetic resonance imaging (MRI) was helpful in the diagnosis.

Case report

A 47-year-old woman, gravida 0, was referred to our hospital with suspected uterine and ovarian tumors, and chief complaints of sudden pain in the right lower abdomen and fever (38.5 °C). She had experienced abdominal distention for several years, with rapid growth over the last 6 months. The patient presented with a giant, soft abdominal mass associated with tenderness in the right lower quadrant (Fig. 1a), and a protruding navel, partially bluish-purple in coloration and involving a subcutaneous hard nodule, measuring 3 × 4 cm (Fig. 1b). The nodule was painless, and the patient had no history of umbilical bleeding or discharge. She had irregular menstrual periods and oligomenorrhea, and felt no dysmenorrhea or menorrhagia. There was no history of medical or surgical diseases.

The white blood cell count was 10,540/ml, hemoglobin was 12.5 g/dl, and C-reactive protein level was 4.32 mg/dl. Level of carbohydrate antigen (CA)125 was slightly elevated, but other tumor markers were within normal ranges: CA125, 58.28 U/ml; CA19-9, 12.25 U/ml; carcinoembryonic antigen (CEA), 0.4 ng/ml; and lactate dehydrogenase (LDH), 206 U/l.

Ultrasonography revealed marked enlargement of the uterus with two solid hypoechoic tumors measuring 14 cm and 11 cm in diameter and suspected to represent leiomyomas, two cystic tumors measuring 12 cm and 7 cm in diameter, and a large tumor containing abundant variable-sized cysts and surrounding the above tumors. Color Doppler ultrasonography showed poor blood flow in these tumors.

MRI revealed an umbilical tumor showing signal hypointensity on T2-weighted imaging and slight signal hyperintensity on fat-suppressed T1-weighting imaging with small foci of slightly signal hyperintensity on T2-weighted imaging, signal hyperintensity on diffusion-weighted imaging and fat-suppressed T1-weighting imaging, and no enhancement with contrast-enhanced MRI. MRI also revealed extended endometrium and multiple large tumors within the uterine body, comprising solid tumors regarded as typical leiomyoma, cystic tumors, and markedly degenerated tumors with cystic and edematous changes showing heterogeneously high signal intensity on T1- and T2-weighted imaging. These uterine tumors were poorly enhanced after injection of gadolinium chelate (Fig. 2). MRI also showed a right ovarian cystic tumor without solid part, measuring 6 cm, containing fluid showing signal hyperintensity on T1-weighted imaging, signal hypointensity on T2-weighted imaging, and poor enhancement. Computed tomography (CT) showed no findings suggestive of apparent lymph node involvement or metastatic disease.

The patient underwent simple hysterectomy, bilateral salpingo-oophorectomy and resection of the umbilical tumor. The cut surface of the nodule within the subcutaneous tissue of the umbilicus was grayish-white with scattered hemorrhagic spots. Rapid intraoperative pathological diagnosis of the umbilical nodule indicated endometriosis. Intraoperative findings showed necrotic cystic tumor of the right ovary

* Corresponding author at: Department of Obstetrics and Gynecology, Faculty of Medicine, University of Yamanashi, 1110 Simokato, Chuo-city, Yamanashi 409-3898, Japan. Fax: +81 55 2738719.
E-mail address: msasaki@yamanashi.ac.jp (M. Omori).

http://dx.doi.org/10.1016/j.gynor.2014.04.004

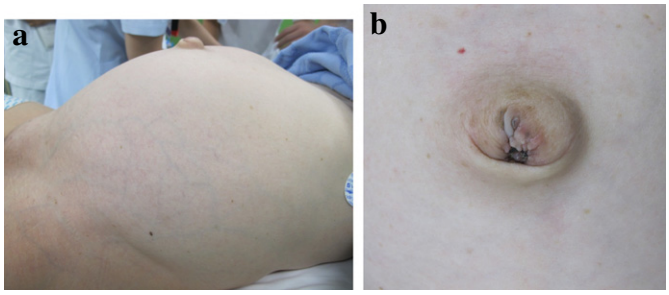

Fig. 1. a) The patient presented with a protruding navel and a giant, soft abdominal mass derived from multiple leiomyomas with marked hydropic or cystic degeneration. b) The protruding navel was partially bluish-purple, and involved a subcutaneous hard nodule measuring 3 × 4 cm.

with 1080° torsion of the pedicle, and a smooth-surfaced, huge uterus with protrusions of variable-sized cystic tumors and firm solid tumors. The removed uterus weighed 9.6 kg, measuring 35 × 25 cm. No evidence of intrapelvic endometriosis was identified.

Histological findings of the umbilical tumor showed a glandular arrangement lined by columnar cells surrounded by stroma of spindle cells, corresponding to endometrial tissue, with scattered intraglandular hemorrhage and marked proliferation of smooth muscle cells and fibroblasts (Fig. 3a). Examination of the uterus showed variable-sized, well-circumscribed tumors with disorderly arrangement of the smooth muscle fascicles, different degrees of hydropic, cystic, and hyaline degeneration, and edematous changes, without any nuclear atypia or mitotic changes (Fig. 3b). The right ovarian tumor showed severely necrotic changes with hemorrhage, and contained bloody fluid. Postoperative diagnosis was endometriosis of the umbilicus, uterine leiomyomas with hydropic and cystic degeneration, and right ovarian cyst with ischemic change. No evidence of malignancy was seen. Postoperative course was uneventful.

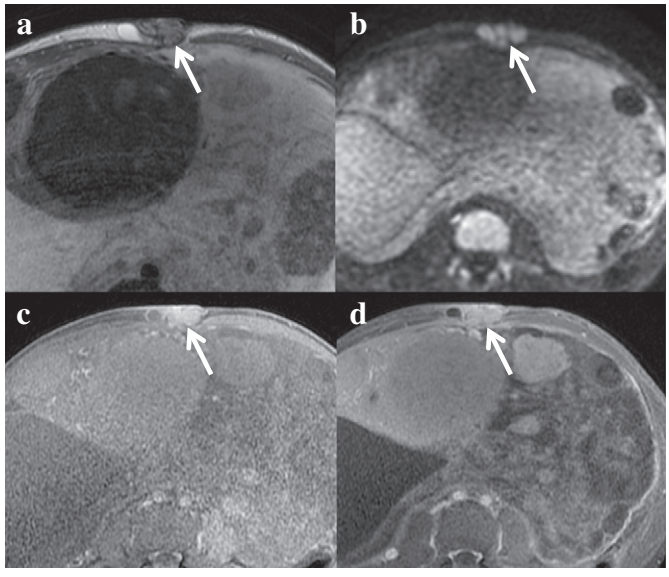

Fig. 2. Axial magnetic resonance (MR) images. a) T2-weighted imaging. b) Diffusion-weighted imaging (b = 1000). c) Fat-suppressed T1-weighted imaging. d) Gadolinium-enhanced T1-weighted imaging. The umbilical tumor (arrow) shows low signal intensity on T2-weighted imaging and slightly high signal intensity on fat-suppressed T1-weighting images with small foci of slight signal hyperintensity on T2-weighted imaging, signal hyperintensity on diffusion-weighted imaging, and non-enhancement with contrast-enhanced MRI. MRI reveals multiple large different tumors in the uterine body comprising solid tumors regarded as typical leiomyoma, cystic tumors, and extensively degenerated tumors with cystic and edematous changes showing heterogeneous signal hyperintensity on T1- and T2-weighted imaging and poor enhancement.

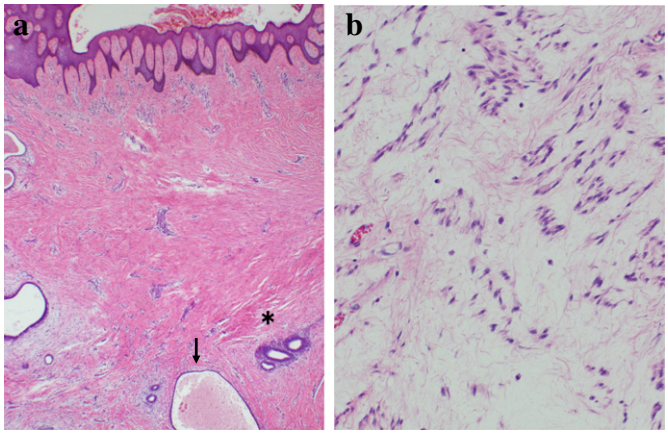

Fig. 3. Histological findings. a) Umbilical endometriosis, showing a glandular arrangement lined by columnar cells surrounded by stroma of spindle cells, corresponding to endometrial tissue (asterisk), with scattered intraglandular hemorrhage (arrow) and marked proliferation of smooth muscle cells and fibroblasts (hematoxylin and eosin, H&E, ×5). b) The leiomyoma with marked hydropic degeneration. Most of the smooth muscle cells were replaced by hydropic fibrous tissue, with only cords of residual, bland, spindle-shaped cells remaining (H&E, ×10).

Discussion

Cutaneous endometriosis is relatively uncommon. The prevalence is reportedly less than 5.5% of all cases of endometriosis, and its occurrence has recently been increasing (Kyamidis et al., 2011). In 70% of cases, cutaneous endometriosis appears in surgical scars such as from cesarean sections, laparoscopy, hysterectomy, or episiotomy, and the remaining 30% appear spontaneously, most commonly on the umbilicus, followed by the inguinal region (Kyamidis et al., 2011; Agarwal and Fong, 2008; Dessy et al., 2008). The pathogenetic mechanisms of cutaneous endometriosis have been speculated to involve iatrogenic implantation or hematogenous or lymphogenous metastasis of endometrial tissue (Latcher, 1953; Kyamidis et al., 2011; Agarwal and Fong, 2008; Dessy et al., 2008). Agarwal and Fong noted that spontaneous cutaneous endometriosis was associated with more severe pelvic disease than scar endometriosis (Agarwal and Fong, 2008).

Umbilical endometriosis presents as a rubbery or firm nodule, and the size varies from several millimeters to 6 cm. Clinical symptoms typically include pain, bleeding, and swelling correlated with the menstrual cycle, but some patients are asymptomatic (Kyamidis et al., 2011; Agarwal and Fong, 2008; Dessy et al., 2008). The pathology is mostly associated with pelvic endometriosis, with complaints of dysmenorrhea, dyspareunia, or defecation pain (Kyamidis et al., 2011). Umbilical endometriosis can be suspected based on the clinical presentation, but the diagnosis should be confirmed by histological examination. Differentiation from malignancy such as SMJN is essential.

Few reports have described MRI findings for umbilical endometriosis. Yu et al. described two cases and Hartigan and Holloway reported one case of umbilical endometriosis, with signal hypointensity on T1-weighted imaging and signal hypo- or hyperintensity on T2-weighted imaging with 2- to 3-mm signal-hyperintense foci on T1-weighted imaging and fat-suppressed T1-weighted imaging. Areas of signal hyperintensity on T1- and T2-weighted imaging were consistent with subacute hemorrhage containing extracellular methemoglobin, and areas of signal hypointensity on T2-weighted imaging were consistent with hemosiderin and fibrotic tissue (Kyamidis et al., 2011; Yu et al., 1994; Hartigan and Holloway, 2005). The present case also showed findings on MRI corresponding to histological findings of thickened fibrotic tissue containing scattered intraglandular hemorrhage. Yu et al. noted that MRI is helpful to delineate the size and locations of lesions and to exclude the possibility of intraabdominal extension. MRI may be more useful in detecting pelvic endometriosis than differential diagnosis of the umbilical nodule.

The treatment of cutaneous endometriosis involves en-bloc surgical excision with wide margins. Treatment using pharmacotherapies such as gonadotropin-releasing hormone agonist (GnRHa) or contraceptive pill alone is considered insufficient, but the medication can be effective for the relief of symptoms derived from pelvic endometriosis (Kyamidis et al., 2011; Agarwal and Fong, 2008). Recurrence is uncommon if complete excision is performed. However, malignant transformation has been reported in 0.3–1% of cutaneous endometriosis, particularly in patients with longstanding or recurrent endometriosis. Common histological subtypes include endometrioid carcinoma, clear cell carcinoma, adenosarcoma, and serous adenocarcinoma (Kyamidis et al., 2011; Agarwal and Fong, 2008).

The present case was initially suspected to involve malignant uterine tumor and its umbilical metastasis, but the findings on ultrasonography and MRI demonstrated a high likelihood of benign uterine tumor. As uterine leiomyomas enlarge, they can outgrow their blood supply, resulting in various types of degeneration, such as hyaline (>60% of cases), cystic (about 4%), myxoid, or red degeneration (Ueda et al., 1999). Reports of leiomyoma with marked cystic degeneration have been relatively uncommon (Aydin et al., 2013). The present case was uncommon in that it exhibited umbilical endometriosis, an extremely enlarged uterus due to marked hydropic and cystic degeneration of the leiomyomas, and torsion of the pedicle of the right ovarian cyst that had caused sudden pain in the right lower abdomen. The correlations of the pathogenesis between these three lesions were unclear. The patient in this case made no complaints of umbilical tumor, symptoms derived from endometriosis, or apparent evidence of intrapelvic endometriosis.

Umbilical nodules should be distinguished from primary carcinoma or metastasis. Nevertheless, umbilical tumors in women involve endometriosis in approximately 30% of cases (Barrow, 1966). Physicians should take into consideration the possibility of umbilical endometriosis in the case of umbilical nodules, particularly among reproductive-age women.

Conflict of interest statement

The authors declare that there are no conflicts of interest.

References

Agarwal, A., Fong, Y.F. (2008). Cutaneous endometriosis. Singap. Med. J. 49, 704–709.
Aydin, C., Eriş, S., Yalçin, Y., Sen, Selim H. (2013). A giant cystic leiomyoma mimicking an ovarian malignancy. Int. J. Surg. Case Rep. 4, 1010–1012.
Barrow, M.V. (1966). Metastatic tumors of the umbilicus. J. Chron. Dis. 19, 1113–1117.
Dessy, L.A., Buccheri, E.M., Chiummariello, S., Gagliardi, D.N., Onesti, M.G. (2008). Umbilical endometriosis, our experience. In Vivo. 22, 811–815.
Dubreuil, A., Dompmartin, A., Barjot, P., et al.(1998). Umbilical metastasis or Sister Mary Joseph's nodule. Int. J. Dermatol. 37 (1), 7–13.
Hartigan, C.M., Holloway, B.J. (2005). Case report: MR imaging features of endometriosis at the umbilicus. Br. J. Radiol. 78, 755–757.
Kyamidis, K., Lora, V., Kanitakis, J. (2011). Spontaneous cutaneous umbilical endometriosis: report of a new case with immunohistochemical study and literature review. Dermatol. Online J. 17 (7), 5.
Latcher, J.W. (1953). Endometriosis of the umbilicus. Am. J. Obstet. Gynecol. 66, 161–168.
Ueda, H., Togashi, K., Konishi, I., Kataoka, M.L., Koyama, T., Fujiwara, T., Kobayashi, H., Fujii, S., Konishi, J. (1999). Unusual appearances of uterine leiomyomas: MR imaging findings and their histopathologic backgrounds. Radiographics 19, S131–S145.
Yu, C.Y., Perez-Reyes, M., Brown, J.J., Borrello, J.A. (1994). MR appearance of umbilical endometriosis. J. Comput. Assist. Tomogr. 18, 269–271.